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*Veterinary
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Animal and
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Bovine Spongiform Encephalopathy

Bovine spongiform encephalopathy (BSE) is a chronic degenerative disease affecting the central nervous systems of cattle. The disease was first diagnosed in 1986 in Great Britain. The British sometimes call BSE "mad cow disease," a term that could be confused with rabies in cattle if used in the United States.

BSE has had a substantial impact on the British livestock industry. The disease also has been confirmed in domestic cattle in Ireland, France, Switzerland, and Portugal and in cattle exported from England to Oman, the Falkland Islands, Germany, Denmark, and Canada. The U.S. Department of Agriculture's (USDA) Animal and Plant Health Inspection Service (APHIS) is enforcing import restrictions and is conducting surveillance for BSE to ensure that this serious disease does not become established in the United States.

Clinical Signs

Cattle affected by BSE experience a progressive degeneration of the nervous system. Affected animals may display changes in temperament, such as nervousness or aggression; abnormal posture; incoordination and difficulty in rising; decreased milk production; and loss of body weight despite continued appetite. Affected cattle die. The causative agent of the disease is still unknown, and there is no treatment.

The incubation period (the time from when an animal becomes infected until it first shows disease signs) is thought to be from 2 to 8 years. Following the onset of clinical signs, the animal's condition deteriorates until it either dies or is destroyed. This usually takes from 2 weeks to 6 months. Most cases in England have occurred in dairy cows between 3 and 5 years of age.

Currently, there is no test to detect the disease in a live animal; veterinary pathologists confirm BSE by postmortem histologic examination of brain tissue. BSE is so named because of the spongy appearance of the brain tissue of infected cattle when sections are examined under a microscope.

Related Diseases

BSE belongs to a group of related diseases known as the transmissible spongiform encephalopathies, which are all caused by uncharacterized agents that produce spongiform changes in the brain. The group includes scrapie, which affects sheep and goats; transmissible mink encephalopathy; feline spongiform encephalopathy; chronic wasting disease of mule deer and elk; and kuru, Creutzfeldt-Jakob disease, and Gerstmann-Straussler syndrome, three rare diseases in humans. Other cases of spongiform encephalopathies have been reported in Great Britain in kudus, an eland, a nyala, and a gemsbok.

No scientific evidence indicates that BSE can be transmitted from infected cattle to humans through contact or consumption of beef or dairy products. The World Health Organization does not consider BSE to be a human health hazard based on current scientific evidence.

Epidemiology

There are different scientific hypotheses concerning the origins of BSE. One theory is that BSE had existed in undetectable levels in the British cattle population prior to 1986. Another theory stems from epidemiologic data that suggest that BSE in England may have been caused by feeding cattle rendered products from the carcasses of scrapie-infected sheep. The practice of using products such as bonemeal in cattle rations as a source of protein has been common for several decades. Scrapie has a long incubation period—up to 60 months—and has been endemic in Great Britain for centuries. Changes in rendering operations in the early 1980's—particularly the removal of a solvent-extraction process and the elimination of a second steam-heat treatment—may have played a part in the appearance of the disease and the large number of cases that developed.

There is no evidence that BSE spreads from cattle to cattle or from cattle to other species by contact. Researchers have not gathered substantial epidemiologic evidence to determine if vertical transmission of BSE occurs. The BSE agent has been found only in brain tissue and the spinal cord in clinical research. In an experimental study of the disease's biological route of development in cattle, traces of BSE were detected in the

small intestines of calves that had been fed large doses of material from BSE-infected animals.

The extremely small infectious agent responsible for BSE and scrapie, although not completely characterized, has been theoretically classified as a "slow virus," a "prion," or a "virino." This agent is extremely resistant to heat and to normal sterilization processes. It also does not evoke a detectable immune response or inflammatory reaction in host animals.

History

During the period from November 1986, when BSE was first identified as a separate disease entity, until the end of 1988, there were nearly 2,200 cases on about 1,700 farms in England. Newly reported cases are currently occurring at a rate between 700 and 750 per week. As of July 1994, more than 132,000 head of cattle in over 30,000 herds were found to be infected in Great Britain. (There are approximately 10 million cattle and 38 million sheep in that country.)

As a result of BSE, agricultural officials in Great Britain have taken a number of actions, including (1) prohibiting the inclusion of ruminant-derived proteins in all animal rations, (2) prohibiting the consumption of milk and meat from suspect animals by either animals or humans (except for milk from a dam to its calf), (3) stopping human consumption of certain bovine organs, (4) destroying all animals showing signs of BSE and indemnifying the owners, and (5) making BSE a notifiable disease.

USDA Actions in Response to BSE

To prevent BSE from entering the United States, APHIS has restricted the importation of live ruminants and ruminant products from countries where BSE is known to exist. Other products derived from ruminants, such as fetal bovine serum, bonemeal, meat with bonemeal, bloodmeal, offal, fats, and glands, also cannot be imported into the United States from these countries, except under a special permit for scientific research purposes.

In addition to international importation restrictions, APHIS has increased surveillance efforts to detect BSE if it is accidentally introduced into the United States. More than 250 APHIS and State veterinarians specially trained to diagnose foreign animal diseases regularly conduct field investigations of suspicious disease conditions. APHIS veterinary pathologists also have received training from their British counterparts for diagnosing BSE.

These pathologists examine brain tissue from cattle over 2 years of age that show signs of neurological disease. More than 60 veterinary diagnostic laboratories throughout the United States have agreed to forward tissue from such animals to the National Veterinary Services Laboratories in Ames, IA, for histologic examination. To date, more than 1,800 specimens from 42 States have been examined, and no evidence of BSE has been seen.

As part of increased surveillance for BSE, APHIS veterinarians are tracing 499 head of cattle imported from Great Britain between 1981 and 1989 (before the ban on imports went into effect) to check their health status. As of September 1994, 371 of the animals had been accounted for, and no signs of BSE had been found. Efforts continue to trace the remaining cattle.

APHIS leads the interagency effort to coordinate surveillance for BSE. Officials of USDA's Food Safety and Inspection Service (FSIS) notify APHIS of cattle having neurological signs at slaughter. The Centers for Disease Control and Prevention also examine the brains of rabies-negative cattle for BSE.

Assuring Accurate Diagnosis

To guard against a misdiagnosis of BSE, APHIS has established a protocol for a definitive diagnosis. Diagnosis of BSE depends extensively on a carefully conducted histologic examination of the brain in the laboratory. However, to verify an initial positive finding, APHIS is requiring additional diagnostic procedures, together with a thorough analysis of clinical and epidemiologic data. The protocol for BSE diagnosis relies heavily on the experience of scientists at the Central Veterinary Laboratory in Weybridge, England. APHIS neuropathologists performing the microscopic evaluations in the program have been trained at Weybridge.

Getting the Word Out

As part of the increased surveillance activities, APHIS is continuing an education effort to inform U.S. cattle producers and veterinarians about this new disease. Numerous briefings have been held for industry groups. In addition to press releases and factsheets, a British videotape on BSE and an information packet were distributed to all APHIS field offices, State veterinarians, extension veterinarians, colleges of veterinary medicine, and industry groups.

For additional information, contact:
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Telephone: (301) 436-8073.

For information about importing animals, contact:
USDA, APHIS, VS, Import-Export Products Staff; Room 756, Federal Building; 6505 Belcrest Road; Hyattsville, MD 20782.
Telephone: (301) 436-7885.

For information about importing animal products, contact:
USDA, APHIS, VS, Import-Export Animals Staff; Room 764, Federal Building; 6505 Belcrest Road; Hyattsville, MD 20782.
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